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# Pancreatic gastrointestinal stromal tumor: A case report

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ABSTRACT

*INTRODUCTION:* Gastrointestinal stromal tumors (GISTs) are the most common gastrointestinal tract (GIT) tumors of mesenchymal origin. GISTs tend to arise with a higher frequency in the stomach and the small intestine. GISTs that originate from outside of the GIT are defined as extra-gastrointestinal stromal tumors (EGISTs). Among them pancreatic EGISTs are very rare.
*CASE PRESENTATION:* A 30 years old male patient presented with abdominal pain. Triphasic abdominal computed tomography scan with contrast revealed large well defined mass at the pancreatic tail, about 12 × 11.6 cm. Laparoscopic distal pancreatectomy and splenectomy was performed. Postoperative pathological examination revealed positive CD 117 and Dog 1 confirming the diagnosis of EGISTs.
*DISCUSSION:* GIST is a rare mesenchymal tumor. EGISTs arising in the pancreas are extremely rare, about, 5% of EGISTs. Its origin remains controversial. Some authors believe that GISTs and EGISTs arise from the common cell origin of interstitial cells of Cajal. Others suggest that EGISTs are at the beginning, mural GISTs with extensive extramural growth, resulting in later on, loss of their connection with the GIT wall.
*CONCLUSION:* We report a rare case of large pancreatic tail EGIST, which was resected, safely and effectively by laparoscopic approach.



## 1. Introduction

GISTs are rare mesenchymal tumors that can arise in any part of the GIT. Most of these tumors arise in the stomach and small intestine [1]. It should be noted that, GISTs that originate from outside of the GIT, are defined as EGISTs and are usually arise from omentum, mesentery and retroperitoneam, adjacent, but separate from the stomach and the intestine [2]. EGISTs that arise from the pancreas are extremely rare, and only 30 cases have been reported in the literature to date [3].

## 2. Case presentation

At our outpatient clinic, 30 years old male patient presented with abdominal pain of 2 weeks duration associated with anorexia and weight loss. The body mass index of the patient was 26.7 kg/m$^2$ and no associated co-morbidity. Physical examination revealed no specific finding. Routine laboratory investigations were within normal limit.

Abdominal ultrasonography (US) showed large mass 15 × 14 × 12 cm at pancreatic tail. Triphasic abdominal CT with contrast revealed large well defined mass at pancreatic tail, about 12 × 11.6 cm, partially solid/cystic, compressing the pancreatic tissue and the splenic vein was seen stretched on outer surface of the mass (Fig. 1).

## 3. The procedure

Laparoscopic distal pancreatectomy and splenectomy was performed. At laparoscopic assessment, large cystic mass about 10 cm in size, at the tail of the pancreas, adherent to splenic vein but not attached to the surrounding organs as stomach. No ascitis, no hepatic focal lesions and no evidence of peritoneal nodules.

Firstly, opening of the lesser sac along the greater curvature of stomach was done for exposing the pancreatic mass (Fig. 2). Trial of dissection along the mass to do localized resection with safety margin but the mass was adherent to splenic vein. So, the decision was to do distal pancreatectomy and splenectomy.

Double ligation and clipping of splenic artery was performed (Fig. 3). Transection of the pancreas using laparoscopic Endo GIA stapler 60 mm (Fig. 4). Mobilization of the spleen by division of leino-renal and spleno-colic ligaments.

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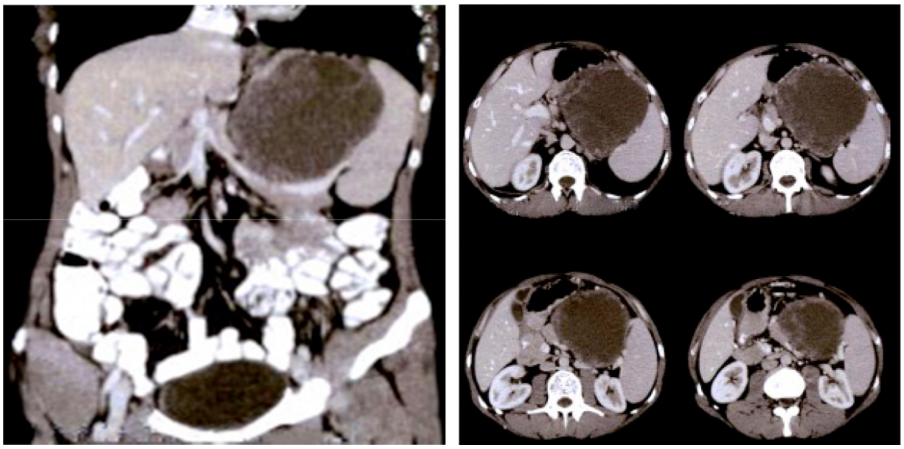

**Fig. 1.** Triphasic abdominal CT with contrast, (coronal and axial view) revealed large well defined, partially solid/cystic mass at pancreatic tail.

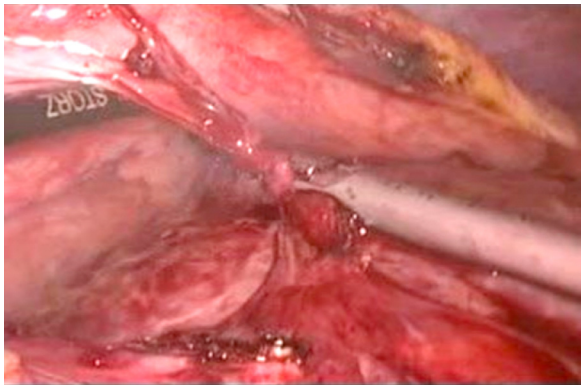

**Fig. 2.** Opening of the lesser sac and exposure of the pancreatic EGIST mass.

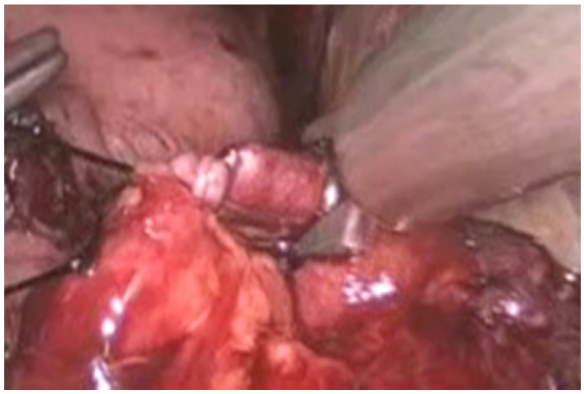

**Fig. 3.** Ligation and clipping of the splenic artery.

Extraction of specimen enbloc through pfennestiel incision was done with good haemostasis and drain at splenic bed. Finally, closure of the incision and abdominal ports.

## 4. Histopathological examination

Macroscopic examination showed spleen measuring 12 × 8 × 3 cm, part of pancreas with attached cystic mass measuring 12 × 11 × 3 cm. The mass was partially cystic, partially solid. Dissection of surrounding fat revealed nine LNs with firm greyish white cut surface.

Microscopically, the tumor was formed of spindle cell proliferation in fasicular pattern with oval shaped nuclei (Fig. 5A&B). The mitotic count was 6 mitoses/50 High Power Fields. Immunohistochemical staining revealed positive immunoreactivity of the tumor cells for CD117 (Fig. 5C) and DOG-1 (Fig. 5D) while smooth muscle actin, S100 protein, CD10 and CD56 were negative. So, the diagnosis was high risk pancreatic GIST with free safety margin and all dissected nine lymph nodes.

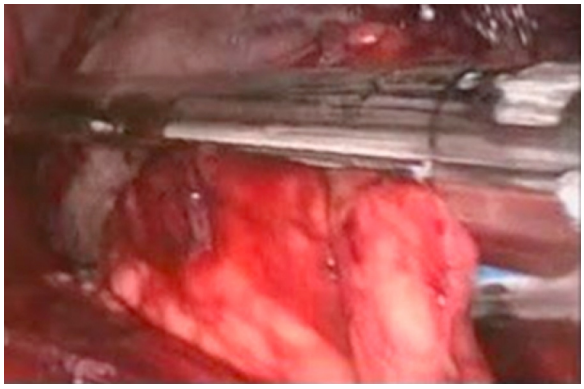

**Fig. 4.** Transection of the pancreas using laparoscopic Endo GIA stapler.

## 5. Post-operative

The patient had oral feeding at 2nd post-operative day (POD). Post-operative laboratory evaluation as blood sugar and serum amylase was within normal range. Drain was removed and the patient was discharged at 3rd POD without any complication.

## 6. Follow up

The patient was referred to oncology department and recieved imitinab therapy in a dose of 400 mg twice daily. There was no local or systemic recurrence detected during the three months follow-up examination.

## 7. Discussion

GISTs are the most common mesenchymal tumors of the GIT [4]. It is believed that these tumors originate from interstitial cells of Cajal (ICCs) [4]. These tumors may present at any site in the GIT where there are ICCs but, the most common locations are stomach, small intestine and colon [5]. EGISTs are stromal tumors that originating from outside the GIT and show no connection to the wall or the peritoneal surface of the GIT [6].

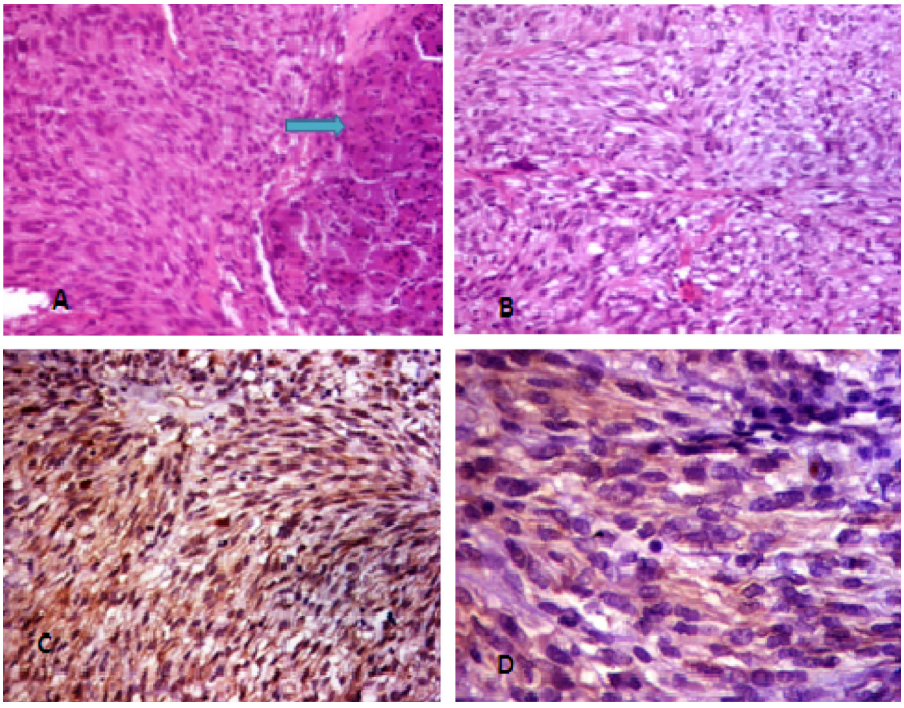


**Fig. 5.** Histomicrographs of GIST. A, The neoplastic proliferation and the adjacent panreaticacini (blue arrow) can be seen (H&E x200). B, The tumor is formed of spindle cell proliferation in fasicular pattern with oval shaped nuclei (H&E x200). C, The neoplastic cells showed positive cytoplasmic staining for CD117 (IHCx200). D, The tumor cells showed positive cytoplasmic staining for DOG1 (IHC x400). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

It is reported that, EGISTs are extremely rare, represent 5%-10% of all GISTs [7,8]. EGISTs are frequently located in the omentum, mesentery or retroperitoneum [9]. EGISTs arising in the pancreas are extremely rare, about, 5% of EGISTs [10].

The clinical presentation of pancreatic EGISTs is variable, depending on the location and size of the tumors in the pancreas [10,11]. The tumor may be discovered accidently [11]. Radiological evaluation of pancreatic tumors is done by abdominal CT, US, and endoscopic US, and aid in determining tumor localization, size, invasion of surrounding organs, and distant metastases; however, most of them are non-diagnostic [12].

The diagnosis of GIST is based mainly on histopathological, immunohistochemical, and molecular features. Histopathologically, GISTs are classified into three main types, spindle, epithelioid, or mixed type. Most pancreatic EGISTs consist of spindle cells [8].

It should be noted that, the most selective immunohistochemical markers of GISTs is the expression of the c-Kit receptor tyrosine kinase (CD117 antigen) in 95% of GISTs. It was reported that EGISTs show similar KIT mutations of typical GISTs suggesting that these tumors have a similar origin [13].

However, the origin of EGISTs still remains controversial. Some authors believe that GISTs and EGISTs arise from the common cell origin of ICCs, which may account for their growth within and outside the GIT .Other explanation suggests that EGISTs are in fact mural GISTs with extensive extramural growth, resulting in lastly, loss of their connection with the GIT [14].

Surgical resection is believed to be the primary preferred line of treatment of pancreatic EGIST [1]. The aim of surgical resection of pancreatic EGIST is to achieve complete resection with clear safety margins [8,15].

Selection the optimal type of surgical resection depends on pancreatic EGIST location. Pancreatecoduodenectomy is the optimal treatment for pancreatic head tumors, while distal pancreatectomy for pancreatic tail tumors [8]. Routine lymph node dissection is not indicated in pancreatic EGIST cases because of rare regional lymph node metastases [11].

Imatinib, which is an inhibitor of the tyrosine kinase activity of C-Kit, has been added as a line of treatment of GISTs [16]. Neoadjuvant and adjuvant therapy with imatinib has been shown to reduce the risk of recurrence and improve the survival [17]. Imatinib can be used as neoadjuvant therapy, for large size GISTs to reduce tumor size, increase the rate of complete resection of the tumor, and help to improve prognosis [8,18].

Laparoscopic distal pancreatectomy is now performed more frequently in the surgical resection of benign and even malignant tumors in the body and tail of the pancreas [19]. Many studies have reported that, Laparoscopic distal pancreatectomy is associated with decreased intraoperative blood loss, shorter hospital stay and less morbidity compared with open distal pancreatectomy [20].

## 8. Conclusion

We report a case of large pancreatic EGIST, which is considered a rare tumor. Laparoscopic distal pancreatectomy can be safely and effectively performed in the curative surgical resection of pancreatic EGIST.

## Consent

Written informed consent was obtained from the patient for publication of this case report.

## Conflict of interest

There is no conflict of interest.

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